Single ventricle with systemic obstruction in early life: comparison of initial pulmonary artery banding versus the Norwood operation

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Received 9 October 2000; received in revised form 2 March 2001; accepted 2 March 2001

Abstract

Objectives: The outcomes of initial pulmonary artery banding (PAB) ± coarctation repair are compared with the Norwood operation in newborns with single ventricle (SV) and systemic obstruction (SO).
Methods: Between January 1987 and July 2000, 22 patients (median age, 12 days) with SV and aortic arch obstruction (AAO), subaortic stenosis (SAS), or both underwent surgery. Two initial surgical approaches were used: PAB ± coarctation repair (group I, seven patients); Norwood type operation (group II, 15 patients).
Results: The overall mortality was 32% (seven of 22 patients). There was no late mortality. The mortality in group I was 43% versus 27% in group II. Recently, there has been no mortality following the Norwood operation in the last eight patients operated since 1995. Of the survivors, nine patients have undergone the Fontan operation and four patients have had the bidirectional Glenn (BDG) with no deaths. There was one repair of supravalvar aortic stenosis at the time of BDG in group II as opposed to eight reinterventions for SAS and/or AAO in four patients in group I (P = 0.01). Conclusions: PAB ± coarctation repair for SV and SO is associated with a high mortality and a high reoperation rate for SAS or recurrent AAO. Although the Norwood operation was also associated with a high mortality early on, it can now be performed with excellent outcome. This improvement, combined with a low reintervention rate for SAS or AAO, suggests that the Norwood operation is likely to emerge as the procedure of choice for SV and SO. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Single ventricle; Systemic obstruction; Aortic arch obstruction; Norwood operation; Pulmonary artery banding

1. Introduction

Most patients with single ventricle (SV) and systemic obstruction (SO) are generally critically ill soon after birth. The combination of SO and the absence of pulmonary obstruction leads to torrential pulmonary overcirculation with rapid development of congestive heart failure. In the presence of aortic arch obstruction (AAO), the lower systemic circulation is maintained by a patent ductus arteriosus (PDA). Its closure after birth leads to acute hypoperfusion of vital organ systems. The natural history of this congenital malformation without surgical treatment is overwhelmingly fatal [1]. At present, the ultimate palliative treatment for this group of patients is the Fontan procedure.

Although remarkable advances in the successful application of the Fontan operation has resulted in tremendous optimism for the long-term outcome of patients with SV, the survival in early life is far from ideal. Indeed, many patients will never become candidates for the Fontan procedure [2–4]. This study reviews our experience with SV and SO and compares the outcome of two philosophically different initial surgical approaches over a period of 14 years.

2. Methods and materials

2.1. Patient population

Between January 1987 and July 2000, 22 consecutive infants were referred to the Montreal Children’s Hospital with the diagnosis of functionally SV and SO. Patients with hypoplastic left heart syndrome were excluded. Of these patients, 12 were males and 10 were females. All patients were managed either by pulmonary artery banding (PAB)
with or without coarctation repair or by a Norwood or Damus–Kaye–Stansel (DKS) type of operation. The median age at initial operation was 12 days (range, 2 days–6.75 months).

2.2. Morphology

The morphological diagnosis was established by a two-dimensional echocardiography, color flow imaging and/or angiography. The diagnoses for the 22 patients entering the comparison analysis are summarized in Table 1. The diagnosis of restrictive bulboventricular foramen (BVF) was established when the size was less than 75% of the aortic annulus or in the presence of the flow gradient across the pathway. Aortic coarctation is present when there is a discrete narrowing at the periductal area, together with the presence of posterior shelf. The aortic arch was considered hypoplastic if its narrowest diameter was less than the weight of the baby in kg plus 1 mm. Right to left shunting across the PDA suggested AAO. Recurrent aortic coarctation is defined on the basis of a pressure gradient over 20 mmHg in the absence of PDA. Subaortic stenosis (SAS) is defined by the presence of morphological obstruction or the presence of a pressure gradient across the subaortic structures. A pressure gradient may not be present despite the presence of obstruction during initial presentation with unobstructed pulmonary artery and when the circulation is ductal-dependent.

2.3. Surgical management

All patients included in this study were operated in the neonatal period, with the exception of one patient who was referred late. This patient underwent a modified Norwood operation at 6 months of age. Instead of a systemic–pulmonary shunt, a bidirectional Glenn (BDG) was performed with success. For the purpose of analysis, this patient was considered in the Norwood management group. Two philosophically different initial surgical approaches were used in the remaining patients. In group B (n = 7), PAB alone was performed in two patients and PAB with coarctation repair was performed in the remaining five patients. In group S (n = 15), a Norwood operation was performed in 13 patients and a DKS type of operation was used in the other two patients. The Norwood operations involved the use of pulmonary homograft patch aortoplasty of the aortic arch and the ascending aorta opened down to the level of the transected pulmonary artery. A 3–4 mm Gore-Tex tube was used to construct a modified Blalock–Taussig or central shunt. Since July 1999, all Norwood or DKS operations in our institution have been performed without deep hypothermic circulatory arrest (DHCA) [5].

2.4. Follow-up

Patients’ records were reviewed retrospectively. Survivors were followed by their referring cardiologists. The status of all patients was inquired by telephone contact of the referring cardiologists or family. Recurrences of AAO, SAS or other anatomical obstructions were noted. Reoperation for relief of SO and the progression to BDG and the Fontan operation were also noted. Cardiac catheterization was routinely performed prior to Fontan operation or in the presence of signs of SO requiring further interventions. The median follow-up was 8 years (range, 7 months–13 years).

2.5. Statistical analysis

The mortality rate was analyzed by Chi-square and Fisher Exact tests between the two groups undergoing different initial surgical managements. Standard statistical methods for analysis of rates were also used to compare the rates of reintervention for SO between the two cohorts. A P
value less than 0.05 was considered to be statistically significant.

3. Results

3.1. Mortality

The overall surgical mortality was 32% (7/22). In patients treated with PAB ± coarctation repair (group B), the mortality rate was 43% (3/7) versus 27% (4/15) in the Norwood/DKS type procedure (group S; P = NS). No late mortality occurred during the period of follow-up. In addition, there was no mortality in group S since 1995 (P = 0.077). Furthermore, there have been no deaths among 11 patients treated by any approach since 1995, as opposed to a 58.3% mortality (7/12) among the 12 patients treated before 1995.

3.2. Progress

Among the four survivors in group B, two patients had further progressed to a BDG at the time of this review and the other two patients had successfully completed the Fontan operation. However, two of these patients required additional systemic–pulmonary shunts through the course of progression.

Among the 11 survivors in group S, five have progressed to a BDG and six patients have successfully completed the Fontan operation. No death occurred during the second and third stage procedures or at late follow-up. All surviving patients are currently doing well.

3.3. Incidence of recurrent SO

Four patients in group B required reintervention to address the new onset or recurrent SAS or AAO versus one patient in group S who developed supravalvar stenosis which was corrected at the time of BDG (P = 0.01 by the Fisher Exact test). In addition, the difference in rate of reintervention was significantly higher among the four group B surviving patients who required eight reinterventions versus one in group S (P = 0.003). The rate of reintervention was 15.3/100 patient years in group B versus 1.1/100 patient years in group S.

4. Discussion

The true incidence of SO associated with functionally SV is difficult to determine because of the extreme heterogeneity of this group of patients. It ranges between 45 and 50% among patients with functional SV and excessive pulmonary blood flow [1,6]. Most of the patients have ductal-dependent systemic circulation to the lower body and they become critically ill as the PDA gradually closes soon after birth, unless resuscitated aggressively. The natural history of this congenital malformation without surgical treatment is overwhelmingly fatal. In a series of 13 patients with SV, AAO, without SAS, who were not treated surgically because of complex anatomy or prohibitive associated medical conditions, 12 died in infancy and the remaining child died at 3.5 years after birth [1]. When there was SV, AAO and SAS at presentation, all of six patients died in infancy. Even with surgical treatment, the survival of neonates with SV and coarctation of the aorta remains poor. Among 435 neonates with coarctation reported by the Congenital Heart Surgeons Society Study between 1990 and 1992, 32 patients had SV [7]. At the conclusion of this relatively short study period, only ten patients survived, resulting in a mortality of 69%. This underscores the difficult surgical challenge among patients with this congenital malformation that optimal surgical therapy remains an elusive goal.

We have previously summarized the goals of initial surgical therapy for SV with SO [8]. Of the highest priority, the establishment of unobstructed systemic circulation is crucial as it serves to maintain good systolic and diastolic function of the SV. Preservation of competent atrioventricular and semilunar valve function will contribute to a good long-term outcome. Particularly important is the achievement of adequate, but not excessive pulmonary blood flow to prevent the development of pulmonary vascular obstructive disease. The prevention of pulmonary venous obstruction and the avoidance of pulmonary artery stenosis complete the goals of surgical management.

Several surgical approaches have been used for the management of patients with SV and SO. However, the choice of procedure for the management of this challenging malformation remains controversial. PAB and coarctation repair had been used the longest and also represented our procedure of choice during the early period of the study. The Norwood operation and the palliative arterial switch operation (ASO) represent new additions to the surgical armamentarium.

4.1. PAB and coarctation repair

The combined procedures of PAB and coarctation repair via a left thoracotomy had been the traditional approach for the management of patient with SV and AAO. This approach has been advocated for the critically ill neonate as a simple first-step palliation that avoids the use of cardiopulmonary bypass (CPB) or DHCA. The PAB serves to decrease the volume overload on the SV, while protecting the pulmonary vascular bed from developing obstructive disease. The potential drawback of PAB and aortic arch repair without the use of CPB includes incomplete relief of AAO when approached via a left thoracotomy. Tubular hypoplasia of the aortic arch has been reported to be as high as 93% among patients with complex intracardiac lesions [9]. Techniques such as subclavian flap angioplasty, coarctectomy, and even extended end-to-end anastomosis, may not be sufficient to adequately address the tubal hypoplasia.
of the arch. More importantly, the majority of these patients managed initially with PAB and aortic arch repair will ultimately develop SAS within the first year of life which requires further intervention, often coupled with the intermediate stage procedure before the Fontan operation. Freedom et al. noted an 84.4% incidence of SAS in patients with SV and discordant ventricular arterial connection physiology treated initially with PAB [10]. Others have also confirmed a similar development of subaortic obstruction after PAB, which is a risk factor for mortality after the Fontan operation [11,12]. Among the 18 patients with double-inlet left ventricle, transposition of great arteries (TGA) and AAO described by Webber et al. [13], one patient underwent a neonatal Norwood procedure and died. The remaining 17 patients underwent aortic arch repair in addition to PAB. One early death resulted in this series. However, among the 16 survivors, 15 patients required relief of SAS with three subsequent deaths. Jensen et al. [14] described a 26 patient cohort with SV physiology. Among them, eight patients had associated AAO and underwent PAB and aortic arch repair. Essentially, all these patients developed SAS over time, which required the DKS procedure to relieve the obstruction with one death. A second patient died after a Fontan operation with very high subaortic gradient preoperatively. Although the remaining six patients have successfully progressed towards the ultimate palliative procedure, four of these six patients had significant residual subaortic gradients, potentially affecting their long-term outcome. The proponents of PAB have suggested that the poor outcome, as well as the development of SAS, with this approach are caused by the excessively long duration of PAB. Therefore, vigorous surveillance by echocardiography is recommended to detect the development of SAS so that early intervention can be implemented. Excellent early results have been reported with this PAB approach [15,16], although the series are equally small and follow-up relatively short. Among the 15 neonates with SV and AAO from the University of California at Los Angeles experience, all patients were treated with primary PAB and relief of AAO as the initial surgical management via left thoracotomy [17]. Thirteen patients were alive at the end of the follow-up period. However, seven patients subsequently required a DKS procedure to relieve the subaortic obstruction at a median age of 4 months. Subsequently, seven patients had progressed to bidirectional cavopulmonary anastomoses and six patients underwent the Fontan procedure with one takedown to a BDG. Furthermore, two other patients have been deemed not suitable for a Fontan operation. Although improvement of survival has been reported by these recent reports, SAS remains a significant problem with this approach which requires vigorous surveillance and aggressive intervention during a long-term follow-up before definitive palliation is achieved. The current report further substantiates this propensity in PAB group. In our experience, recurrent or new onset SO occurred in 3/4 survivors in group B. A fourth patient died with persistent AAO after PAB. Our results showed that there is an approximately 15 times increase in the rate of reintervention/100 patient years associated with initial PA banding ± relief of AAO. The presence of preexisting BVF obstruction in a neonate with SV and AAO has generally been considered a contraindication to PAB. However, in the setting of a critically ill neonate with congestive heart failure and a large PDA, it may be difficult to assess the true pressure gradient across the foramen. The presence of coarctation or AAO should, in itself, suggest the potential of SO at the BVF level, as suggested by Mattiau et al. [18]. It is therefore not surprising to find a high incidence of SAS in the setting of SV with AAO because of the morphological substrate inherent to these defects. Donofrio et al. further suggested that the volume unloading of the ventricle caused by PAB might alter the ventricular geometry, resulting in acute diminution of the BVF size and worsening the SAS [19]. The placement of a PAB in this setting will naturally result in the creation of a doubly obstructed heart, accelerating the process of myocardial hypertrophy, ischemia and fibrosis, with eventual myocardial failure. Inherent complications associated with PAB have been well documented. Proximal placement of the band can potentially result in subsequent damage to the pulmonary valve, causing pulmonary insufficiency. Distal migration of the band may lead to branch pulmonary artery stenosis. These complications may prohibit the patient from further progressing in the Fontan pathway.

4.2. Norwood and DKS type operations

The principle of the Norwood or DKS type of operation uses the main pulmonary artery as an additional egress from the heart into the systemic circulation, and the pulmonary circulation is based on shunt physiology. This early aggressive approach has been advocated by several groups as a primary procedure for the neonates with SV and AAO [8,20–21]. It bypasses multiple levels of obstruction. As the two outflow tracts originating from the SV are now contributing to the systemic circulation, the presence or subsequent development of native SAS becomes irrelevant. Our experience supports this hypothesis and has resulted in a significantly reduced need for multiple reinterventions due to new onset or recurrent SAS or AAO. This should contribute to a better Fontan candidate in the long-term. The main drawback with the Norwood or the DKS type of operation has been the increased risk of a complex operation during the neonatal period while the patient is usually critically ill. The use of CPB and DHCA would increase the immediate risk of the operation. The presence of a shunt physiology further complicates the post-operative management. Jacobs et al. have reported the experience with the Norwood operation for lesions other than hypoplastic left heart syndrome and revealed an overall mortality of 28%, with 13 of 18 survivors all deemed to be good Fontans [20]. Others have reported a similar success with the use of the Norwood...
procedure as a first stage palliation with an equivalent rate of progression into the subsequent Fontan pathway [21]. Ilbawi et al. have also been advocates of a primary pulmonary-to-systemic anastomosis in patients with SV with AAO because of the consistent appearance of severe SAS [22]. They noted that patients with SV and SAS, with or without AAO, had significantly lower ventricular mass and mass-to-volume ratios when they were treated initially with pulmonary artery to aortic anastomosis versus PAB followed by subsequent relief of SAS. In addition, the former group had good outcomes after the Fontan operation, whereas the group palliated with PAB and coarctation repair had a high perioperative mortality and required takedown of the Fontan because of low cardiac output and high venous pressure.

4.3. Palliative ASO

The experience with the palliative ASO is limited to a small number of cases [23,24]. The premise of this approach in patients with SV and AAO is a very high likelihood for the development of SAS. The performance of a palliative ASO transfers the subaortic obstruction into the subpulmonary level. Subsequently, the pulmonary vasculature will be protected naturally by the restrictive BVF, while a harmonious aortic root is reconstructed without the use of prosthetic material. An additional advantage of this surgical option is the avoidance of a systemic to pulmonary shunt. However, this represents a much greater operation, requiring transfers of the coronary ostia and the need for CPB or DHCA. However, the restriction of pulmonary blood flow by the restrictive BVF may be greater than expected because of the creation of an unobstructed neoaortic outflow tract that preferentially promotes flow through the systemic circulation. Therefore, the pulmonary blood flow tends to be unpredictable and there is a potential for additional systemic to pulmonary shunt as the restrictive BVF becomes hemodynamically more significant with time.

4.4. Alternative surgical approaches

Other alternative surgical approaches, which have been used much less frequently, have been described. Simultaneous PAB, coarctation repair and enlargement of the BVF have been reported by Serraf et al. [25], who recommended that this approach should only be limited to the older patients because of the difficulty of exposing the subaortic area in neonates. Another uncommonly used surgical therapy includes the use of an apical left ventricular aortic conduit. However, its use in the neonatal population has been limited by the inevitable change of the synthetic valved conduit, as well as the potential damage to the ventricular function. Orthotopic heart transplantation is also used as a form of surgical therapy in those patients with very poor function of the SV precluding any palliative approach. The long-term result is expected to be comparable with other patients requiring heart transplantations.

At the Montreal Children’s Hospital, we have used essentially two of these surgical approaches since 1987, and they are compared in the current report. This retrospective analysis of our surgical experience over a 14 year period came out heavily in favor of the Norwood/DKS approach. The absence of surgical mortality since 1995 coincided with the decision to use the Norwood operation for SV with AAO. Of importance is the significantly higher reintervention rate for recurrent or residual SO with the PAB approach. This undoubtedly has a negative impact on the preservation of diastolic ventricular function as the patient awaits the Fontan operation.

The current analysis has several limitations. The number of patients were accumulated over a long period of time, during which, there were rapid changes in the surgical and medical management of these patients. The study was non-randomized and retrospective, so that the observational data may have been confounded by other unmeasured factors. Due to the small number of patients with SV and SO seen by individual institutions, it is likely that definitive proof of the optimal surgical approach may require the performance of a prospective multi-institutional study.

In conclusion, the traditional approach of the PAB and aortic arch repair for SV and SO has been associated with a high mortality and a high reoperation rate for SAS and AAO. The use of either a Norwood or DKS technique has neutralized the hemodynamic significance of SAS or AAO. The Norwood operation can now be performed with low mortality without circulatory arrest and can have excellent mid-term results. This improvement, combined with a low reintervention rate for SO, suggests that the Norwood or DKS type operation will optimize the performance of the Fontan candidate and is likely to emerge as the procedure of choice for SV and SO.

References

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Appendix A. Conference discussion

Dr V. Alexi-Meskhishvili (Berlin, Germany): When you decide to perform some kind of operation of the systemic obstruction, please define what is the systemic obstruction. What criteria are you using for systemic obstruction? Is it pressure gradient or is it BVF orifice ratio to the aortic ring ratio, or some other, because it’s a very practical issue?

Dr Tchervenkov: Obviously, the definition of systemic obstruction is very important. These are mostly patients who had a ductal-dependent systemic circulation. The vast majority had aortic arch obstruction with coarctation and hypoplasia of the aortic arch, ± subaortic stenosis. There were only four patients in this series who had subaortic stenosis with a normal aortic arch. It is very difficult to elicit gradients when you have an open ductus and when the single ventricle can eject into the pulmonary circulation at the same time. So, our criteria were ductal dependence of the systemic circulation or in its absence, if the bulboventricular foramen was less than about 80% of the aortic root, or if there was an anatomic substrate for the potential of subaortic stenosis.

There is obviously a gray area, but I believe that we should be overly aggressive rather than wait for the obstruction to arise. There are groups that subscribe to initial pulmonary artery banding and coarctation repair, such as Hillel Laks in LA. This is followed by an aggressive follow-up of these patients, and conversion to a Glenn with a DKS operation as soon as there is evidence for subaortic obstruction. That is one way of doing things, but we prefer to be more aggressive.

Dr M. Magalhaes (Lisbon, Portugal): Would you be more precise about this banding problem. If you’ve got a single ventricle situation and sooner or later you’re going to go into either a Damus–Kaye or a Norwood procedure, can you comment on any advantage on doing the banding and trying to control the patient initially with it?

Dr Tchervenkov: I think the study illustrates the problems with banding, at least in our center. The patients who have died after the banding approach have done so, I believe, because of a doubly-obstructed heart. Now, you have to take all of this a little bit with a grain of salt. I think this study does not definitely prove that the only way to go is the Norwood operation. For that, we need a multi-institutional prospective randomized study with a much larger number of patients. The problem is that most centers have a very small number of patients with this diagnosis. So, I think for a conclusive proof, you need that kind of a study. I hope I answered your question.

Dr A. Corno (Lausanne, Switzerland): You may have already answered with the previous question, but if you are convinced that the mortality is better without the banding, and if you know that with the banding, you create the prerequisite for failure of a univentricular type of repair, what is your criteria now to use a banding again, in which patient?

Dr Tchervenkov: We published a treatment algorithm in the Advances in Cardiac Surgery in 1999 which shows our current preference. If you have no anatomic substrate for subaortic stenosis and you don’t have subaortic stenosis, we subscribe to a pulmonary artery band, followed by an early Glenn. For any other situation, as you can see in this algorithm, we advocate the Norwood operation or a DKS procedure. It would be very unfortunate to put a PA band, and later on to have to do a DKS and to find that the pulmonary valve has been damaged. That has happened to me on a couple of occasions. So again, this is a treatment algorithm we propose, but we need a prospective multi-institutional study for definitive proof.

Dr P. Vouhe (Paris, France): Maybe we could ask Dr Daenen to comment on this paper because, very recently, he published a paper on the same subject and advocated the use of pulmonary banding.

Dr W. Daenen (Leuven, Belgium): Actually, last year, we presented 14 cases, and of these 14, seven had very severe arch obstruction coarctation; in these seven patients, we did a coarctation arch repair together with a banding. You have to place your banding very accurately, and thereafter,
you have to follow your patients very, very carefully, because in this subset of patients, they very often develop subaortic stenosis very soon. The earliest we had to go to a Glenn and Damus-Kaye-Stansel was after 6 weeks. With this policy, we were able to have survival in all seven patients; five of those went on to a successful complete Fontan operation.

_Dr Tchervenkov_: I think that’s excellent. There are many ways to skin the cat. However, the experience that you just alluded to and Hillel Laks’ experience is that the vast majority of these patients will develop subaortic stenosis. What we are saying is, why do things in multiple stages when we can address the potential or the presence of systemic obstruction with one operation, at the initial operation. Obviously, this approach will have to withstand the test of time, but we are not the only group that advocates this approach.